CHAPTER 61

Examination of the Motor System: Approach to Weakness

KEY TEACHING POINTS

- Neuromuscular weakness can have any of four causes: upper motor neuron disease (central weakness), lower motor neuron disease (peripheral weakness), neuromuscular junction disorders, and muscle disease. Each of these disorders is associated with distinct physical signs, neuroanatomy, and etiologies.
- The combination of both upper and lower motor neuron findings indicates disease of the spinal cord, the only anatomic location where both segments reside.
- Special tests such as pronator drift, the forearm rolling test, and the finger tapping test accurately detect contralateral cerebral hemispheric disease even when muscle power is largely preserved.
- In patients with stroke, the presence of aphasia or conjugate eye deviation accurately localizes the stroke to the anterior circulation (i.e., in the distribution of the internal carotid arteries). In contrast, a stroke affecting the posterior circulation (i.e., distribution of the vertebral and basilar arteries) is more likely if there is Horner syndrome, crossed sensory or motor findings, nystagmus, heterotropia, ataxia, or hemianopia.

THE MOTOR EXAMINATION

Examination of the muscles includes inspection (for atrophy, hypertrophy, fasciculations, and tremor), percussion (for myotonia), palpation (for abnormal tone), full flexion and extension of the elbows and knees (for abnormal tone and nonneurologic restrictions to movement, such as contractures or joint disease), and tests of muscle strength.

I. MUSCLE STRENGTH

A. DEFINITIONS

Paralysis refers to a loss of power of any degree, from mild weakness to complete loss. The suffixes **plegia** and **paresis** also indicate paralysis (e.g., hemiplegia), although the term *paresis* is usually used to indicate incomplete paralysis. **Tetraparesis**

indicates weakness of all four limbs (specialists in spinal cord disorders prefer this term over *quadriparesis*). **Paraparesis** indicates weakness of both legs; **hemiparesis**, weakness of an arm and leg on one side of the body; and **monoparesis**, weakness of just one arm or leg.

B. THE FINDINGS

1. TECHNIQUE

The clinician tests a single muscle at a time by asking the patient to contract the muscle strongly while the clinician tries to resist any movement. Unilateral weakness is recognized by comparing the muscle to its companion on the opposite side; bilateral weakness is recognized by comparing the strength to some standard recalled from clinical experience. The clinician grades the muscle's strength according to a 6-point system (0 through 5), as described later. (See the section on Grading Muscle Strength.)

In patients with weakness, the clinician should systematically test all the muscles from head to foot, paying particular attention to which muscles are weak, whether proximal and distal muscles of a limb differ in strength, and whether the weakness of a monoparetic limb involves only muscles from a single spinal segment or peripheral nerve (see Chapter 64). An excellent, inexpensive handbook describes the proper technique for testing all of the important muscles of the arms and legs.[1]

Testing muscles by resisting their action, however, tends to overlook significant weakness at the hips and knees, where powerful antigravity muscles can easily overcome the physician's resistance even when significant weakness is present.[2] A better way to test these muscles is to use the patient's own body weight as the load the muscle must lift. For example, quadriceps weakness on the symptomatic leg becomes more apparent when the patient is asked to rise from a chair rather than by manually resisting the patient's attempt to extend the knee.[3] Another method measures the time required by the patient to rise from a chair and then sit down again 10 times. Patients without weakness accomplish this in less than 20 to 25 seconds (<20 seconds if 50 years old and <25 seconds if 75 years old). If patients require more time, proximal weakness of the legs is present unless there is an alternative explanation, such as joint or bone disease.[4]

2. GRADING MUSCLE STRENGTH

Muscle strength is graded using a conventional scale developed by the British Medical Research Council (MRC) during World War II (Table 61.1).[1] This scale, which is universally used, has one important drawback: it assigns a disproportionate amount of a muscle's power to grade 4 strength. For example, the biceps muscle uses just 2% of its full power to overcome gravity (i.e., grade 3 strength), meaning that almost 98% of the remaining range of power is grade 4.[5] Because of this drawback,

TABLE 61.1 Grading Muscle Strength

Grade	Finding
0	No contraction
1	Flicker or trace of contraction
2	Active movement with gravity eliminated
3	Active movement against gravity
4	Active movement against gravity and resistance
5	Normal power

Based upon reference 1.

many neurologists subdivide grade 4 into three more grades: 4 minus (i.e., barely moves against resistance), 4, and 4 plus (i.e., almost full power).

3. SPECIAL TESTS FOR UNILATERAL CEREBRAL LESIONS

In patients with cerebral lesions, measures of muscle power alone often underestimate the size of the lesion and the patient's functional disability. Special tests have been developed as more sensitive tests of motor function in these patients: upper limb drift (**pronator drift**), the **forearm rolling test** (and its variants—index finger test, little finger test, and thumb rolling test),[6,7] and the **rapid finger tapping** and **foot tapping** tests (Fig. 61.1).

C. CLINICAL SIGNIFICANCE

See the section on Approach to Weakness, later.

II. ATROPHY AND HYPERTROPHY

A. ATROPHY

1. DEFINITION

Atrophy describes muscles that are emaciated or wasted.

2. TECHNIQUE

Atrophy is detected during inspection of the muscle. Examples are (1) an abnormally flat thenar eminence when viewed from the side (e.g., cervical radiculopathy or carpal tunnel syndrome), (2) missing shadows on the anterior neck from atrophic sternocleidomastoid muscles (e.g., syringomyelia), or (3) metacarpal bones appearing unusually prominent on the back of the hand, from atrophic intrinsic muscles (e.g., polyneuropathy).

Significant asymmetry of the circumference of the arms or legs indicates atrophy of the smaller side (or edema of the other side). In normal persons, the difference in calf circumference between the right and left sides is less than 1 cm in 90% and less than 1.5 cm in 100% (measured 10 cm below the tibial tuberosity).[11]

3. CLINICAL SIGNIFICANCE

Atrophy is a feature of lower motor neuron disease* and muscle disuse (especially from adjacent joint disease or trauma). In patients with sciatica, the finding of ipsilateral calf wasting (i.e., maximum circumference at least 1 cm less than that of the contralateral side) accurately indicates lumbosacral nerve compression from disc herniation (LR = 5.2, see Chapter 64).

B. HYPERTROPHY

Hypertrophy describes abnormal enlargement of a muscle. Bilateral calf hypertrophy is a typical feature of some muscular dystrophies, although it is found in a wide variety of neuromuscular diseases.[13]

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*In the evaluation of weakness, a fundamental distinction is the separation of upper motor neuron lesions (i.e., located in the cerebral cortex, brainstem, or descending motor pathways of the spinal cord) from lower motor neuron lesions (i.e., located in the peripheral nerves and anterior horn cells of the spinal cord). William Gowers first distinguished the upper and lower motor segments in his 1888 *Manual of Diseases of the Nervous System*.[12] See Fig. 61.2 and the section titled "Approach to Weakness" later in this chapter.

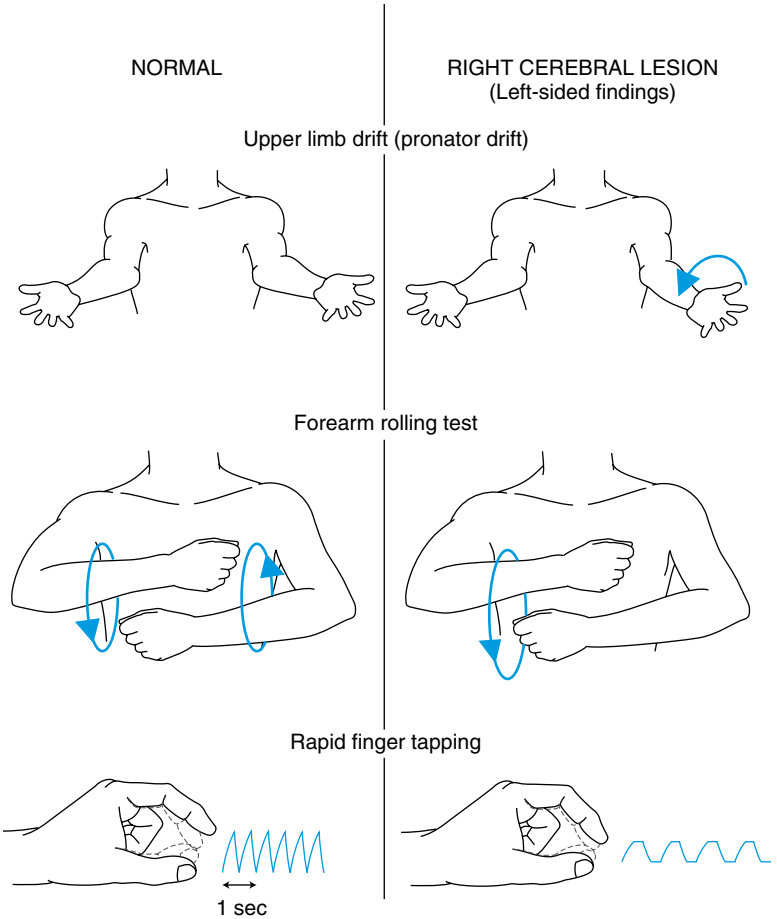


FIG. 61.1 SPECIAL TESTS FOR UNILATERAL CEREBRAL LESIONS. The depicted patient has a right cerebral lesion with left-sided findings during three different tests: **(1) Upper limb drift** (**pronator drift,** *top row*). The patient stretches out both arms directly in front of him or her with palms upright (i.e., forearms supinated) and closes his or her eyes. This position is held for 45 seconds.[8,9] The arm contralateral to the hemispheric lesion drifts downward and pronates. **(2) Forearm rolling test** (*middle row*).[6] The patient bends each elbow and places both forearms parallel to each other. He or she then rotates the forearms about each other in a rapid rolling motion for 5 to 10 seconds in each direction. In the abnormal response, the forearm contralateral to the lesion is held still while the other arm "orbits" around it. **(3) Rapid finger tapping** (*bottom row*). The patient rapidly taps the thumb and index finger repeatedly at a speed of about two taps per second. In normal persons the movement has an even rhythm and large amplitude. Hemispheric lesions cause the contralateral finger and thumb to tap more slowly and with diminished amplitude, as if the finger and thumb are sticking together.[8] The index finger rolling test and little finger rolling test are similar to the forearm rolling test (each index finger or little finger is rotated about the other for 5 seconds in both directions). In the foot tapping test, the seated patient taps one forefoot at a time for 10 seconds on the floor, as fast as possible, while the heel maintains contact with the floor. A discrepancy of more than five taps between the left and right foot indicates cerebral disease contralateral to the slower foot.[10]

III. FASCICULATIONS

A. DEFINITION

Fasciculations are involuntary rapid muscle twitches that are too weak to move a limb but are easily felt by patients and seen or palpated by clinicians.[14] Most healthy people experience fasciculations at some time, especially in the eyelid muscles.

B. CLINICAL SIGNIFICANCE

Isolated fasciculations without other neurologic findings are benign.[15] When accompanied by weakness or atrophy, however, fasciculations indicate lower motor neuron disease, usually of the anterior horn cell or proximal peripheral nerve. Tongue fasciculations occur in up to one-third of patients with amyotrophic lateral sclerosis.[16] (See the section on Approach to Weakness, later.)

IV. MUSCLE TONE

Muscle tone refers to the involuntary muscle tension perceived by the clinician on repeatedly flexing and extending one of the patient's limbs. Such an assessment of muscle tone assumes that the patient is relaxed and that there are no bone or joint limitations to movement. Muscle tone may be increased (e.g., spasticity, rigidity, or paratonia) or diminished (flaccidity).

A. INCREASED MUSCLE TONE

1. SPASTICITY

A. DEFINITION

Spasticity is increased muscle tone that develops in patients with upper motor neuron lesions.[17] The increased muscle tone of spasticity has three characteristics, as follows: (1) **Velocity-dependence.** The amount of muscle tone depends on the velocity of movement: the more rapid the movement, the greater the resistance; the slower the movement, the less the resistance. (2) **Differing tone in flexors and extensors.** The tone in the flexors and extensors of a limb is not balanced, which commonly causes characteristic resting postures of that limb (see later). (3) **Associated weakness.** The muscle with spasticity is also weak. If left untreated, muscles shortened by spasticity may eventually develop fixed contractures.

B. CHARACTERISTIC POSTURES

In spasticity, an imbalance in flexor and extensor tone commonly causes abnormal postures of the resting limb. In hemiplegia, for example, there is excess tone in the *flexors* of the arms and *extensors* of the legs, causing the arm and hand to be fixed against the chest, flexed and internally rotated, and the leg to extend with the foot pointed (see Fig. 7.4 in Chapter 7).[18] In contrast, some patients with complete spinal cord lesions have excess tone in the *flexors* of the legs, which causes the legs to flex up onto the abdomen (**paraplegia in flexion**).[19†]

† These hemiplegic and paraplegic postures recall the neurologic development of normal infants. Paraplegia in flexion resembles the initial posture of babies, with their legs flexed against their chests. After descending pathways from the brainstem mature enough to overcome the spinal reflexes responsible for the flexed position, the infant is eventually able to extend the legs and stand (resembling the extensor tone of hemiplegia). After cerebral connections mature enough to provide fine motor control, the infant becomes able to walk. Damage to the cerebral hemispheres (e.g., stroke) disrupts this fine motor control and uncovers the extensor posture; damage to the spinal cord (e.g., severe multiple sclerosis or complete spinal cord transection) removes all supraspinal inputs, unopposing the original flexed posture of the legs.[17]

C. CLASP-KNIFE PHENOMENON

Up to half of patients with spasticity have the **clasp-knife phenomenon**, a finding usually observed in the knee extensors and less often in the elbow flexors.[18,20] To elicit this phenomenon, the clinician extends the patient's knee using a constant velocity, but as the patient's knee nears full extension, the muscle tone of the quadriceps muscles increases dramatically and completes the movement, just as the blade of a pocket knife opens under the influence of its spring.[12] The clasp-knife phenomenon occurs because a muscle's tone is dependent on the muscle's length, the tone diminishing with stretching and increasing with shortening.

D. RELATIONSHIP OF SPASTICITY TO WEAKNESS

Although spasticity is a sign of upper motor neuron disease, its severity correlates poorly with the degree of weakness or hyperreflexia. Patients with slowly developing lesions of the cerebral hemisphere usually develop spasticity and weakness in concert.[21] Patients with strokes or spinal cord injuries, in contrast, develop immediate weakness and flaccidity, spasticity appearing only days to weeks later.[18] Some elderly patients with large strokes have persistent **flaccid hemiplegia**, in which the paralyzed muscles never develop increased muscle tone despite being hyperreflexic.[21]

2. RIGIDITY

A. DEFINITION

Rigidity is increased muscle tension with three characteristic features: **(1) No velocity-dependence.** The resistance to movement is the same with slow and rapid movements. **(2) Flexor and extensor tone is the same. (3) No associated weakness**. Patients with rigidity lack the clasp-knife phenomenon.[17] **Cogwheel rigidity** refers to rigidity that intermittently gives way, as if the patient's limb were the lever pulling over a ratchet (see Chapter 66).

B. DISTINGUISHING SPASTICITY FROM RIGIDITY

Most clinicians distinguish spasticity from rigidity by repeatedly extending and flexing the patient's limbs and observing the characteristics already noted. In the 1950s, Wartenberg‡ introduced a simple bedside test to assess motor tone and distinguish spasticity from rigidity.[22,23] In this test, the patient is seated on the edge of the examining table, which is open underneath to allow the legs to swing back and forth unobstructed. The clinician lifts both feet to extend the knees, instructs the patient to relax, and then releases the legs. The normal lower limb swings back and forth 6 or 7 times, smoothly and regularly in a perfect sagittal plane. In patients with spasticity, the limbs drop with normal velocity, but their movements are jerky and fall out of the sagittal plane, with the great toe tracing zigzags or ellipses. In patients with rigidity, the swinging time and velocity are significantly reduced, resulting in a total of only one or two swings. Others have confirmed Wartenberg's findings.[24]

C. CLINICAL SIGNIFICANCE

Rigidity is a common finding of extrapyramidal disease, the most common example of which is Parkinson disease (see Chapter 66).

‡Robert Wartenberg, who wrote many popular neurology textbooks in the 1950s, was an ardent opponent of eponyms and called his test the *test for pendulousness of the legs*.

3. PARATONIA

A. DEFINITION

Paratonia is excess muscle tension that is *not present at rest* but develops when the patient's limb *contacts* another object, as if such contact made the patient unable to relax. There are two forms: **oppositional paratonia (gegenhalten)** and **facilitatory paratonia (mitgehen)**. In patients with oppositional paratonia, the clinician feels a stiffening of the limb with every applied movement; but unlike rigidity, the stiffening depends entirely on contact and its force is proportional and opposite to the examiner's movements. Patients with facilitatory paratonia, in contrast, actively aid movements guided by the examiner.

B. TECHNIQUE

One simple test of facilitatory paratonia is to take the arm of the seated patient and bend the elbow back and forth three times, from full flexion to 90 degrees of extension. The clinician then releases the arm in the patient's lap and scores any further movement, 0 being no movement, 4 full flexion or more, and 1 to 3 intermediate movements.[25]

C. CLINICAL SIGNIFICANCE

Both oppositional and facilitatory paratonia are associated with extensive frontal lobe disease and often appear in dementing illnesses.[25] Among patients with dementia, the severity of oppositional or facilitatory paratonia (including the score for the paratonia test described in the previous section) correlates inversely with the score on the Folstein mini-mental state examination ($r = -0.5$ to -0.7, $p < 0.05$).[25]

B. DECREASED MUSCLE TONE: HYPOTONIA (FLACCIDITY)

1. DEFINITION

Hypotonia refers to reduced or absent muscle tension.

2. TECHNIQUE

There are many ways to detect the flaccid muscle: the limb feels "like a rag doll," the muscles feel soft and flabby, the outstretched arm when tapped demonstrates wider than normal excursions, or the knee jerks are abnormally pendular. The original definition of abnormally pendular knee jerks—more than three back-and-forth swings of the patient's leg during testing of the knee jerk—should be revised, however, because many normal individuals have this finding.[26]

3. CLINICAL SIGNIFICANCE

Hypotonia is a feature of lower motor neuron disease and cerebellar disease.

4. PATHOGENESIS

There is some evidence that "normal" muscle tone actually consists of tiny muscle contractions that help the clinician to move the extremity (even though the patient is trying to relax).[27] The clinician perceives reduced muscle tension in hypotonic limbs because these contractions are absent.

V. MUSCLE PERCUSSION

Striking the muscle with a reflex hammer may elicit two abnormal findings, percussion myotonia and myoedema.

A. PERCUSSION MYOTONIA

1. THE FINDING

Percussion myotonia is a prolonged muscle contraction that lasts several seconds and causes a sustained dimple to appear on the skin. Percussion myotonia of the

thenar eminence may actually draw the thumb into sustained opposition to the fingers.

2. CLINICAL SIGNIFICANCE

Percussion myotonia is a feature of some myotonic syndromes, such as myotonia congenita and myotonic dystrophy.[28]

B. MYOEDEMA

1. THE FINDING

Myoedema is a focal mounding of muscle at the point of percussion lasting for seconds. Unlike myotonia, myoedema causes a lump instead of a dimple, and the lump may be oriented crosswise or diagonal to the direction of muscle fibers.[29] An instructive video of the finding is available.[30]

Graves and Stokes originally described myoedema in 1830.

2. CLINICAL SIGNIFICANCE

Myoedema is a normal physiologic response and does not necessarily indicate disease.[31] Its historical association with undernourished patients simply reflects the ease with which the response appears when there is no intervening subcutaneous fat.[29,31] Myoedema is frequently described in hypothyroidism, where the finding correlates with the severity of disease. In one study, myoedema was elicited in 13% of patients with mild hypothyrodism (thyroid-stimulating hormone [TSH] 50 to 100 mIU/L), 29% of those with moderate disease (TSH 100 to 150 mIU/L), and 62% of those with severe disease (TSH >150 mIU/L).[32]

APPROACH TO WEAKNESS

I. CAUSE OF WEAKNESS

Neuromuscular weakness has four principal causes: (1) upper motor neuron disease (*pyramidal tract disease* or *central weakness*), (2) lower motor neuron disease (*denervation disease* or *peripheral weakness*), (3) neuromuscular junction disorders, and (4) muscle disease. Each disorder is associated with distinct physical signs (Table 61.2), neuroanatomy (see Fig. 61.2), and etiologies (Table 61.3).

Most patients with weakness have disorders involving lesions of the upper and lower motor neurons. Clinicians should consider muscle disease in any patient with *symmetric* weakness of the *proximal* muscles of the arms and legs (sometimes associated with muscle pain, dysphagia, and weakness of the neck muscles). Disorders of the neuromuscular junction should be considered in patients whose weakness *varies* during the day or who have *ptosis* or *diplopia*. Associated abnormalities of sensation, tone, or reflexes of the weak limb exclude muscle or neuromuscular junction disease and argue instead for lesions of the upper or lower motor neurons.

II. THE FINDINGS

A. UPPER VERSUS LOWER MOTOR NEURON LESIONS

Both upper and lower motor neuron weakness tend to affect *distal* muscles in either a symmetric or asymmetric pattern. The bedside observations that distinguish these

TABLE 61.2 Differential Diagnosis of Weakness*

Location of Lesion	MOTOR EXAMINATION: Muscle Tone	MOTOR EXAMINATION: Atrophy or Fasciculations?	SENSORY FINDINGS	MUSCLE STRETCH REFLEXES	OTHER FINDINGS
Upper motor neuron	Spasticity	No	Sometimes	Increased	Babinski sign
Lower motor neuron	Hypotonia	Yes	Usually†	Decreased/absent	
Neuromuscular junction	Normal or hypotonia	No	No	Normal/decreased	Ptosis, diplopia
Muscle	Normal	No‡	No	Normal/decreased	Myotonia

*These characteristics are *specific* but not *sensitive* and thus are helpful when *present*, not when absent. See text.
†Sensory findings are in the distribution of spinal segment, plexus, or peripheral nerve. See Chapter 64.
‡Atrophy may be a late finding.

two disorders are other neurologic findings in the weak limb, certain localizing signs of upper motor neuron disease, the Babinski sign, and the type of weakness produced.

1. ASSOCIATED FINDINGS IN THE WEAK LIMB (SEE TABLE 61.2)

Spasticity and hyperreflexia indicate central weakness; hypotonia, atrophy, fasciculations, and absent muscle stretch reflexes indicate peripheral weakness. In patients with central weakness, sensory abnormalities vary from the isolated loss of cortical sensations in the distal limb to dense loss of all sensation throughout the limb; if sensory abnormalities occur in peripheral weakness, they follow the distribution of spinal segments or peripheral nerves (see Chapter 64).

2. LOCALIZING SIGNS OF UPPER MOTOR NEURON WEAKNESS

The upper motor neuron pathway extends from the cerebral cortex down through the spinal cord (see Fig. 61.2), traveling in tight quarters with central neurons innervating other structures. Consequently, in addition to producing central weakness, lesions along this pathway cause characteristic additional physical signs (Table 61.4) that confirm that the weakness is of the central type and pinpoint its location.

3. BABINSKI SIGN

The Babinski sign (see Chapter 63) indicates central weakness. In the positive response, the great toe moves upward after a scratching stimulus to the sole of the patient's foot.

4. DISTRIBUTION OF WEAKNESS

A. LIMBS AFFECTED

The findings of monoparesis, paraparesis, and tetraparesis are unhelpful by themselves because they may occur with either central or peripheral weakness. Only hemiparesis is specific, indicating a central lesion.

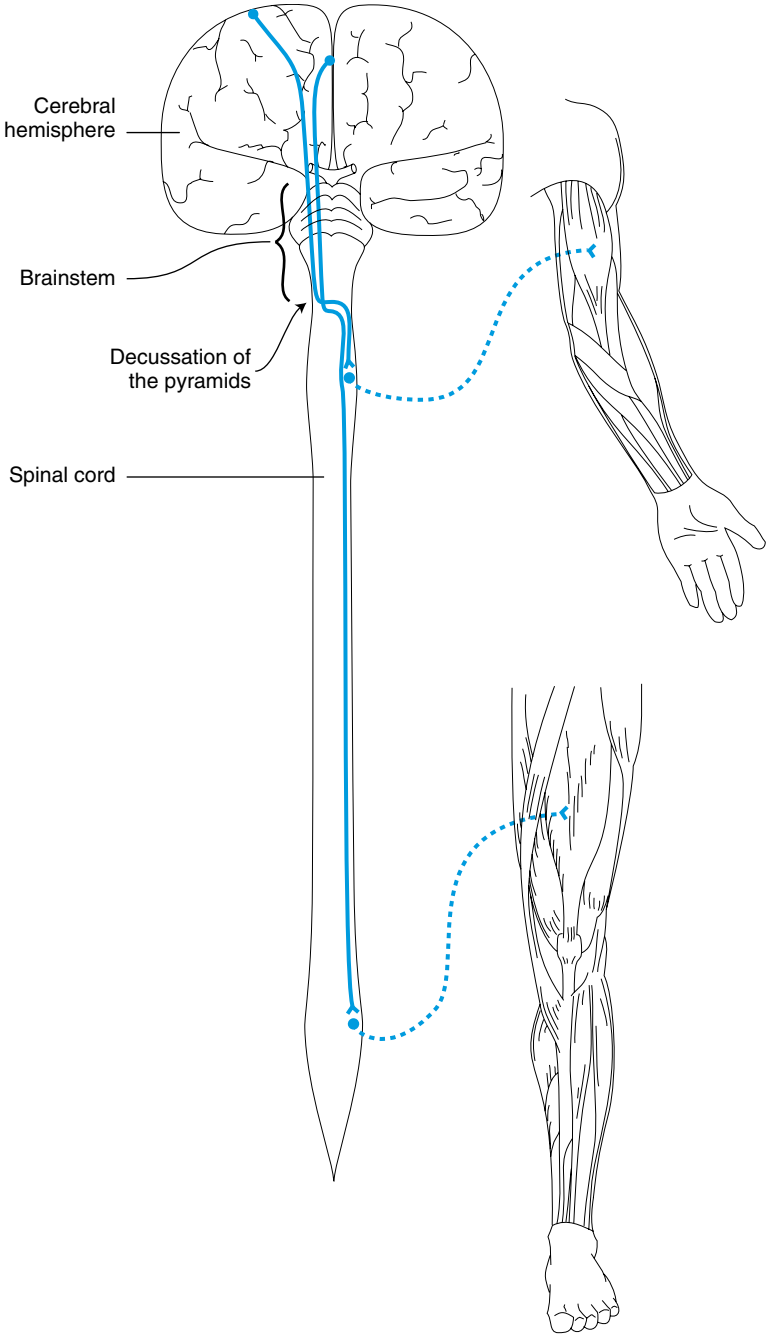


FIG. 61.2 ANATOMY OF UPPER AND LOWER MOTOR NEURONS. The figure illustrates the entire pathway of nerves responsible for movement, from cerebral cortex to muscle. *Upper motor neurons* (*solid line*) extend from the cerebral cortex through the brainstem to the spinal cord. *Lower*

motor neurons (*dotted line*) originate in the spinal cord and travel to muscles within peripheral nerves. Because the upper motor neurons cross to the contralateral side at the border between the brainstem and spinal cord (decussation of the pyramids), weakness of the upper motor neuron type may result from lesions in the *ipsilateral* spinal cord, *contralateral* brainstem, or *contralateral* cerebral hemisphere. Lesions of the spinal cord, where both upper and lower motor neurons reside, may cause weakness of both types: of the *lower motor neuron* type *at* the level of the lesion and of the *upper motor neuron* type in muscles whose peripheral nerves originate *below* the level of the lesion.

B. MOVEMENT VERSUS MUSCLE

Central lesions paralyze *movements*; peripheral lesions paralyze *muscles*. This occurs because neurons from a single area of the cerebral cortex connect with many different spinal cord segments and muscles to accomplish a particular movement. A single muscle has many movements and thus receives information from many different upper segments, all of which converge on the single peripheral nerve traveling to the muscle. A lesion in that nerve therefore obliterates a muscle's entire repertoire of movement; a lesion in an upper segment eliminates only one of many possible movements.[23]

TABLE 61.3 Common Etiologies of Neuromuscular Weakness

Location of Lesion	Common Etiology
Upper motor neuron	Cerebrovascular disease Multiple sclerosis Brain tumor
Lower motor neuron	Polyneuropathy (diabetes, alcoholism) Entrapment neuropathy Trauma
Neuromuscular junction	Myasthenia gravis
Muscle	Drug-induced myopathy Thyroid disease Polymyositis

TABLE 61.4 Localizing Signs in Upper Motor Neuron Weakness

Anatomic Location	Associated Finding
Cerebral hemisphere	Seizures Hemianopia Aphasia (right hemiparesis) Inattention to left body, apraxia (left hemiparesis) Cortical sensory loss* Hyperactive jaw jerk
Brainstem	Crossed motor findings† Contralateral third nerve palsy (midbrain) Contralateral sixth nerve palsy (pons) Sensory loss on contralateral face*
Spinal cord	Sensory level* Pain and temperature sensory loss on contralateral arm and leg* No sensory or motor findings in face Additional lower motor neuron findings (atrophy, fasciculations)

*Chapter 62 describes the different sensory syndromes.
†*Crossed motor findings* refers to unilateral cranial nerve palsy opposite the side of weakness.

One example of this is the contrast between peripheral facial weakness (Bell palsy), which paralyzes all ipsilateral facial movements, and central facial weakness (e.g., from a stroke), which paralyzes voluntary movements but spares emotional ones (e.g., during laughing or crying; see Chapter 60).[33] Another example is the contrast between the peripheral paraparesis of Guillain-Barré syndrome, which paralyzes all leg movements, and the central paraparesis of spinal cord injury, which eliminates volitional movements of the legs but allows the powerful flexor spasms induced by a mild scratching of the patient's foot.[19]

B. THE DIAGNOSTIC PROCESS

1. UPPER MOTOR NEURON WEAKNESS

In patients with upper motor neuron weakness, associated neurologic findings indicate the *level* of the lesion (see Table 61.4); the distribution of weakness indicates the *side* of the lesion. For example, bilateral weakness (paraparesis or tetraparesis) indicates *bilateral* lesions (in the thoracic cord or higher if paraparesis and in the cervical cord or higher if tetraparesis). Monoparesis or hemiparesis indicates a *unilateral* lesion, either in the *contralateral* cerebral hemisphere or brainstem or the *ipsilateral* spinal cord.[§]

Fig. 61.3 illustrates this diagnostic process in the analysis of central weakness. In the first column is the distribution of central weakness for hypothetical patients, which narrows the diagnostic possibilities to a smaller region of the central motor pathway (second column). The associated findings (third column) identify the level of the lesion within that region, thus pinpointing the lesion's location (fourth column).

2. LOWER MOTOR NEURON WEAKNESS

In patients with monoparesis of the lower motor neuron type, the clinician should determine whether the muscles affected are supplied by a single spinal segment (**radiculopathy**), a peripheral nerve (**peripheral neuropathy**), or a combination of the two (**plexopathy**). Further evaluation of these patients is discussed in Chapter 64.

In lower motor neuron weakness, the lesion is always *ipsilateral* to the side of the weakness.

3. COMBINED UPPER AND LOWER MOTOR NEURON WEAKNESS

Combined upper and lower motor neuron findings indicate disease in the spinal cord, the only anatomic location where both segments reside. Common causes are myelopathy and amyotrophic lateral sclerosis.

A. MYELOPATHY

Myelopathy is a term describing a spinal cord lesion confined to a discrete level (e.g., trauma, tumor, disc disease). The lesion causes motor, sensory, and reflex abnormalities *at* the level of the lesion and *below* it. The weakness is of the peripheral type *at* the level of the lesion (from damage to anterior horn cells and spinal roots)** and of the central type *below* the level of the lesion (from damage to the paths of the descending upper motor neuron).

[§] It is the contralateral cerebral hemisphere and brainstem because the descending central motor pathways originate in the contralateral hemisphere, but it is the ipsilateral spinal cord because these pathways cross just below the brainstem (see Fig. 61.2).

** Exceptions to this are lesions at the foramen magnum and C3-C4 level, which sometimes produce atrophy in the hands.[34]

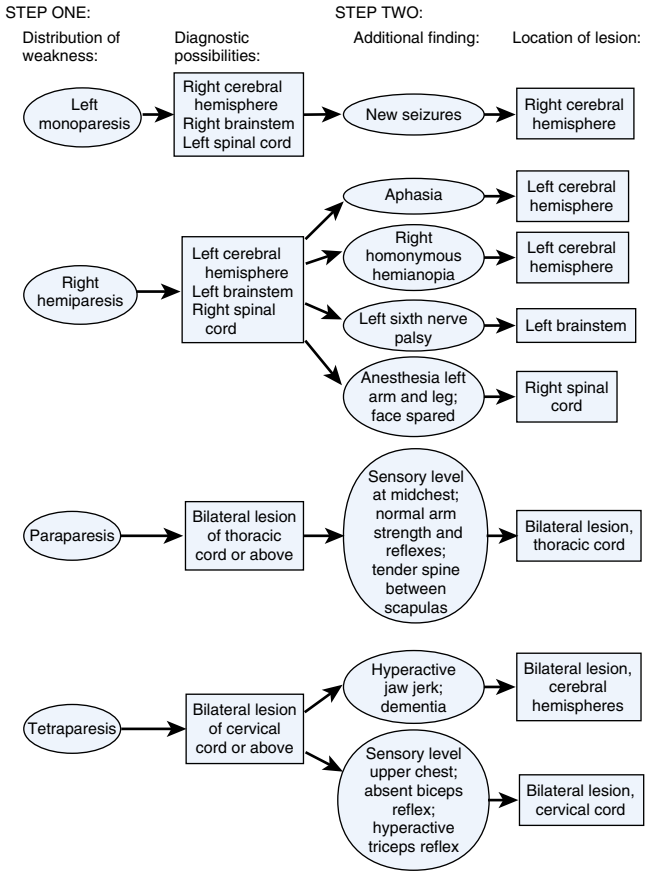


FIG. 61.3 DIAGNOSTIC APPROACH TO UPPER MOTOR NEURON WEAKNESS. The figure illustrates the sequential steps in identifying the location of an upper motor neuron lesion. See the text.

To identify the level of the lesion one must know which spinal segments innervate which muscle. Table 61.5 presents the standardized segmental innervation used internationally by spinal cord specialists (Chapter 64 discusses the derivation of this table). For example, in a patient with a lesion involving the C7 segment of the spinal cord, there is peripheral weakness in the C7 muscles (i.e., atrophy and weakness of the elbow extensors) but central weakness in all the muscles below this level (hyperreflexia and increased tone of the hands, legs, and feet and a positive Babinski sign). The muscles from segments above C7, the biceps and wrist extensors, are normal.[††]

[††] By convention, the neurologic level in spinal cord injury refers to the most caudal level with *normal* function, rather than the first level with abnormal function.[36] The motor level for this hypothetical patient is C6.

TABLE 61.5 Segmental Innervation of Muscles*

Spinal Level	Muscles
ARM	
C5	Elbow flexors (biceps, brachialis)
C6	Wrist extensors (extensor carpi radialis longus and brevis)
C7	Elbow extensors (triceps)
C8	Finger flexors (flexor digitorum profundus of middle finger)
T1	Little finger abductors (abductor digiti minimi)
LEG	
L2	Hip flexors (iliopsoas)
L3	Knee extensors (quadriceps)
L4	Ankle dorsiflexors (tibialis anterior)
L5	Long toe extensors (extensor hallucis longus)
S1	Ankle plantarflexors (gastrocnemius, soleus)

*Most muscles are innervated by nerves from more than one spinal root. This table, based on reference 35, simplifies this innervation to standardize the description of spinal cord injury. A more thorough description of segmental innervation of muscle appears in Figs. 64.1 and 64.6 of Chapter 64.

B. AMYOTROPHIC LATERAL SCLEROSIS

Amyotrophic lateral sclerosis is a degenerative disorder of descending motor tracts and motor nuclei of the spinal cord. The disorder causes both lower motor neuron findings (atrophy, fasciculations) and upper motor neuron findings (hyperreflexia). About half of these patients have a Babinski response.[16] The disease may start in the arms (44%), legs (37%), or bulbar muscles (causing tongue fasciculations, change in voice, and difficulty swallowing) (19%).[16] There are no sensory findings.

Amyotrophic lateral sclerosis and cervical myelopathy are commonly confused at the bedside, even by experienced neurologists.[37] In patients with both upper and lower motor neuron signs, findings that increase the probability of amyotrophic lateral sclerosis are (1) prominent fasciculations, (2) absent sensory findings, and (3) signs of lower motor neuron degeneration affecting more than one level of the spinal cord simultaneously.[38,39‡‡]

III. CLINICAL SIGNIFICANCE

The clinical significance of the motor examination cannot be tested in the conventional manner of this book because bedside criteria alone are sufficient to diagnose many causes of weakness (e.g., cerebrovascular disease, amyotrophic lateral sclerosis, and peripheral nerve injuries are routinely diagnosed by bedside criteria; see Chapter 1).

Nonetheless, several investigations allow a few conclusions.

A. CLINICAL SYNDROMES ARE OFTEN INCOMPLETE

Most studies show that the full lower motor or upper motor neuron syndromes, as depicted in Table 61.2, are often incomplete. In upper motor neuron weakness, up to 25% of patients lack exaggerated reflexes[40,41] and the absence of spasticity

‡‡ The four spinal cord levels are bulbar (jaw, face, tongue, larynx), cervical (neck, arm, hand, diaphragm), thoracic (back, abdomen), and lumbosacral (back, abdomen, leg, foot).

is common, especially in acute lesions (see earlier discussion). Similarly, in many cases of lower motor weakness, the nerve affected does not even innervate a clinical reflex (e.g., L5 radiculopathy, median or ulnar neuropathy); the reflexes of the limbs are thus preserved. Therefore, in the evaluation of weak patients, the *absence* of spasticity or hyperreflexia does *not* argue against the presence of upper motor neuron disease, nor does the *absence* of hypotonicity or hyporeflexia argue against the presence lower motor neuron disease.

On the other hand, the *presence* of abnormal reflexes is very helpful: in one study of patients with weakness, 87% had abnormal reflexes, and in every case areflexia correctly predicted lower motor neuron disease and hyperreflexia correctly predicted upper motor neuron disease.[42]

The fact that syndromes are often incomplete emphasizes the importance of the complete neurologic examination. For example, in a patient with weakness of the fingertips in whom the absence of sensory or reflex changes prevents classification of the weakness as peripheral or central (using the criteria of Table 61.2), the discovery of any additional neurologic finding from Table 61.4 indicates that the lesion is central and pinpoints its location precisely.

B. PROXIMAL WEAKNESS INDICATES MUSCLE DISEASE

If *proximal weakness* is defined as the strength of a limb's proximal muscles being one MRC grade less than that of the distal muscles, proximal weakness is found in 92% of patients with muscle disease.[42] The *absence* of proximal weakness, therefore, decreases the probability of muscle disease.

C. THE SPECIAL TESTS FOR CEREBRAL HEMISPHERIC LESIONS ARE ACCURATE

EBM Box 61.1 presents the diagnostic accuracy of various physical signs for detecting unilateral cerebral hemispheric lesions in patients undergoing computed tomography or magnetic resonance imaging (MRI) of the head. Most of the patients in these studies lacked motor weakness by conventional power testing, and neuroimaging was performed to assess headaches, seizures, or other neurologic symptoms. In these patients, the findings that increase the probability (likelihood ratio, or LR) of *contralateral* cerebral hemispheric lesions the most are a positive forearm rolling test (LR = 15.6), pronator drift (LR = 9.6), Babinski response (LR = 8.5), index finger rolling test (LR = 6), hyperreflexia (LR = 5.3), positive finger tapping test (LR = 4.7), and hemianopia (LR = 4.3). The *absence* of pronator drift (LR = 0.3) diminishes the probability of contralateral cerebral disease.

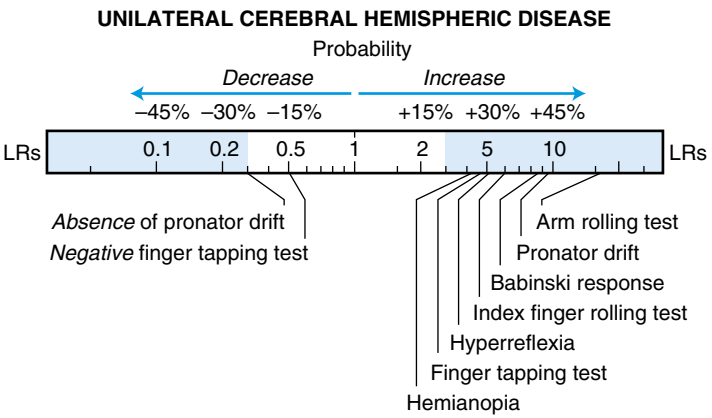
D. ADDITIONAL SIGNS DISTINGUISHING STROKES OF THE ANTERIOR VERSUS POSTERIOR CIRCULATION

Four arteries supply the brain: the right and left internal carotid arteries and the right and left vertebral arteries. The two internal carotid arteries supply most of the cerebral hemispheres (except the posterior occipital lobes) and collectively are called the **anterior circulation.** The two vertebral arteries unite to form the basilar artery; together, these arteries supply the brainstem, cerebellum, and posterior cerebrum (occipital cortex) and are called the **posterior circulation**. Strokes in the distribution of either circulation may produce hemiparesis, but because the anterior and posterior circulations also supply areas of the brain with unique functions, additional telltale findings localize the infarction more accurately. For example, the anterior circulation supplies the areas of the brain controlling language and conjugate eye movements (i.e., movement of both eyes in the same direction, such as to the right or left sides). The posterior circulation, in contrast, supplies areas

EBM BOX 61.1
*Unilateral Cerebral Hemispheric Disease**

Finding (Reference)†	*Sensitivity (%)*	*Specificity (%)*	Likelihood Ratio‡ if Finding Is	
			Present	*Absent*
Cranial Nerves				
Hemianopia[6,42]	22-30	93-98	**4.3**	0.8
Motor Examination				
Pronator drift[6,8,10,43]	22-91	90-98	**9.6**	**0.3**
Arm rolling test[6,8,10,43]	17-87	97-98	**15.6**	0.6
Index finger rolling test[10,43]	33-42	92-98	**6.0**	0.7
Little finger rolling test[10]	7	95	NS	NS
Finger tapping test[6,8,10,43]	16-79	88-98	**4.7**	0.5
Foot tapping test[10,43]	11-23	89-93	NS	NS
Sensory Examination				
Hemisensory disturbance[6]	29	98	NS	0.7
Reflex Examination				
Hyperreflexia[8,43]	11-69	88-95	**5.3**	NS
Babinski response[6,10,43]	9-45	98	**8.5**	NS

*Diagnostic standard: for *unilateral cerebral hemispheric disease*, MRI or computed tomography.
†Definition of findings: for arm rolling test, pronator drift, and finger tapping test, see Fig. 61.1.
‡Likelihood ratio (LR) if finding present = positive LR; LR if finding absent = negative LR.
NS, Not significant.
Click here to access calculator



essential to balance, pupillary function, and alignment of the eyes (i.e., keeping both eyes aligned in the same direction). Also, because descending motor tracts from the brain to the limbs *cross* just below the brainstem, an injury on one side of the *brainstem* (i.e., posterior circulation) may produce *ipsilateral* cranial nerve findings but *contralateral* limb findings (i.e., crossed motor or sensory findings; see Fig. 61.2, Table 61.4, and Fig. 62.2b).

These traditional teachings were confirmed in one study of 1174 patients with strokes, all of whom underwent MRI to localize the injury to the anterior or posterior circulation.[43] In these patients, the following findings increased probability of *anterior* circulation stroke: aphasia (LR = 19.1, EBM Box 61.2) and conjugate gaze palsy (i.e., difficulty moving *both* eyes in an aligned fashion to the right or left) (LR = 3.9). The probability of *posterior* circulation stroke was increased if the following findings were present: Horner syndrome (LR = 72), crossed sensory findings (LR 54.7), crossed motor paresis (LR = 24), nystagmus (LR = 14), heterotropia (i.e., the eyes are not aligned) (LR = 10), ataxia (LR = 5.8), and hemianopia (LR = 3.4). Some findings were diagnostically unhelpful, appearing just as often in anterior circulation stroke as in posterior circulation stroke. These include altered consciousness, hemiparesis, dysarthria, and seizures (LRs not significant or close to the value of 1).

EBM BOX 61.2

Stroke: Anterior Versus Posterior Circulation[*43]

Finding (Reference)[†]	*Sensitivity (%)*	*Specificity (%)*	**Likelihood Ratio[‡] if Finding Is**	
			Present	***Absent***
Detecting Anterior Circulation Stroke				
Aphasia	22	99	**19.1**	0.8
Conjugate gaze palsy	11	97	**3.9**	0.9
Detecting Posterior Circulation Stroke				
Ataxia	32	95	**5.8**	0.7
Horner syndrome	4	100	**72.0**	NS
Hemianopia	4	99	**3.4**	NS
Heterotropia	7	99	**10.0**	NS
Nystagmus	12	99	**14.0**	0.9
Crossed motor paresis	4	100	**24.0**	NS
Crossed sensory findings	3	10	**54.7**	NS

*Diagnostic standard: for *posterior* or *anterior circulation stroke,* localization by MRI.
†Definition of findings: for *ataxia*, see Chapters 7 and 65; for *Horner syndrome*, see Chapter 21; for *hemianopia*, see Chapter 58; for *nystagmus*, see Chapter 65; for *crossed motor paresis* and *crossed sensory findings*, the facial motor or sensory finding is contralateral to the body motor or sensory finding; for *aphasia,* impaired production or comprehension of spoken or written language; for *conjugate gaze palsy*, deviation of both eyes to one side, usually (if cerebral hemispheric stroke) to the side of the lesion and contralateral to the side with weakness.
‡Likelihood ratio (LR) if finding present = positive LR.
NS, Not significant.
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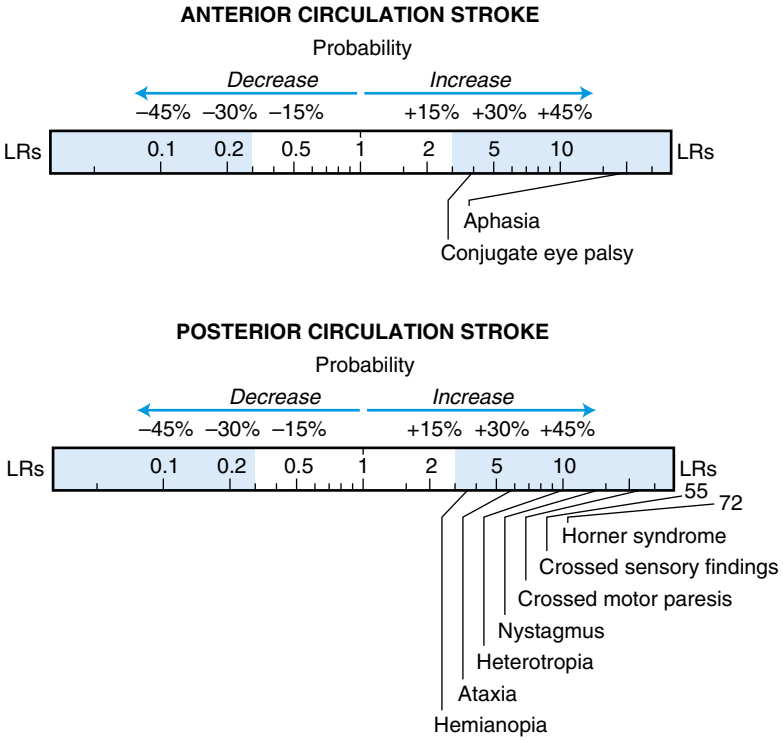
Continued



Importantly, the negative LRs for all of the above diagnostic findings are not significant or close to the value of 1. This means that, even though the *presence* of a particular finding is helpful, its *absence* is not. For example, the *presence* of crossed motor findings is pathognomonic for brainstem infarction (posterior circulation) (LR = 24), but the *absence* of crossed motor findings in a patient with stroke does not at all change the probability of posterior (or anterior) circulation stroke (LR = NS).

E. DIAGNOSIS OF PERIPHERAL NERVE DISORDERS

Chapter 64 discusses the clinical significance of muscle weakness and its localizing value to the diagnosis of peripheral nerve disorders.

The references for this chapter can be found on www.expertconsult.com.

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